

## Huge immature teratoma of the ovary with gliomatosis peritonei in childhood

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### Abstract

Germ cell tumors account for less than 3% of all ovarian cancers. These tumors generally appear in childhood or in those under 30 years of age. Immature ovarian teratoma is the third most frequent germ cell tumor after dysgerminoma and endodermal sinus tumors. These tumors should be distinguished from mature teratomas. Discrimination of malignant and benign tumors depends on the presence of the neuroectodermal components, made up of neural and glial cells. Gliomatosis peritonei is the intraabdominal and particularly peritoneal and omental distribution of the neuroectodermal components, observed very rarely with immature teratoma. Mature teratoma, on the other hand, is even rarer. This report aims to discuss a case of immature teratoma completely filling the abdomen and concomitant omental distribution related gliomatosis peritonei in a 7-year-old child.

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### Introduction

Teratoma tumors of the ovary are the most frequently observed germ cell tumors. Teratomas originate from all three layers of the germ cells and they have many subtypes depending on mature or immature cell content. The common subtypes are mature teratomas (MT), immature teratomas (IT) and monodermal teratomas. Among these, “mature cystic teratoma,” which is also known as a dermoid cyst, is the most frequently observed, with well-known clinical and radiological findings.<sup>1,2</sup> Like MTs, ITs are lesions that may include all three germ cells.<sup>3</sup> ITs are different than MTs for numerous reasons, including: their malignant clinical progressions, being early age lesions (commonly the first 2 decades), being very rare, being unilateral at a high rate, and their including histologically immature or

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embryonic cells.<sup>4,5</sup> ITs (mean: 14-25 cm) typically reach larger sizes than mature cystic teratomas (mean: 7 cm). ITs also have a more solid and less cystic component, include fat and calcification, and can make mass extensions to the neighboring tissues with perforation to the mass wall. Immature teratomas constitute 3% of all ovarian teratomas, 1% of all ovarian cancers, and 20% of malignant germ cell ovarian tumors.<sup>3,6</sup> Mature teratomas are even rarer, and the distribution of the neuroepithelial tissues—particularly in the immature teratomas to the neighboring tissues, such as omentum and peritoneum—is defined as gliomatosis peritonei.<sup>7,8</sup> Distribution of particularly immature neuroepithelial tissues to the neighboring tissues in immature teratomas is defined as metastasis. This metastatic immature neuroepithelial distribution has been staged by Norris et al. according to the modified Robboy and Scully system.<sup>7</sup> Extra-ovarian distributions are very important with regard to prognosis even if they are of microscopic size. However, mature peritoneal or lymphoid distributions do not affect the prognosis negatively and these are accepted as benign structures, since they include mature tissues.<sup>8</sup> Immature tissue-related malignant transformation is very rare in gliomatosis peritonei.<sup>9</sup> Almost all of the dysgerminomas are unilateral ovarian tumors if the 15% possibility of bilateral appearance is excluded.<sup>10,11</sup> There is a 60% chance of early stage diagnosis of this group of tumors seen especially in children. Protective surgery may be planned in patients with progressed tumors as well, since the tumor includes unilateral ovary.<sup>10,11</sup>

In the case of our 7-year-old, a left ovarian mass filled the whole abdomen including both cystic and solid components, we performed a unilateral salphingo-oophorectomy and omentectomy. The pathology report of the case included mature glial implants within the grade 1 immature teratoma and grade 0 omental tissue. The case is taken under follow up.

### **Case Presentation**

A 7-year-old girl was admitted to our clinic with increasing swelling, pain and constipation complaints. In the examination, a full distended abdominal structure with palpable regular and unfixed appearance was observed and small incisions known as “fisset” among the population were observed in all areas of the abdomen’s anterior wall, which are believed to reduce the pain (Figure 1).



**Figure 1: The view of preoperative**

**abdominal distention and applied traditional cuts on the skin called 'fisset'.**

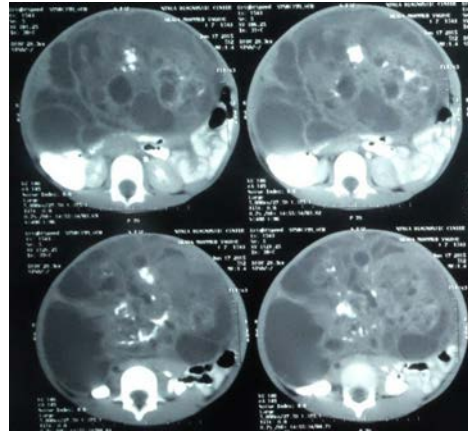
Ultrasonography revealed a 20 x 20 cm. mass that filled the whole abdomen with both solid and cystic components and the origin of which could not be defined precisely. Anatomic structures of the intraabdominal organs could not be clearly visualized in the ultrasonography (Figure 2-USG).



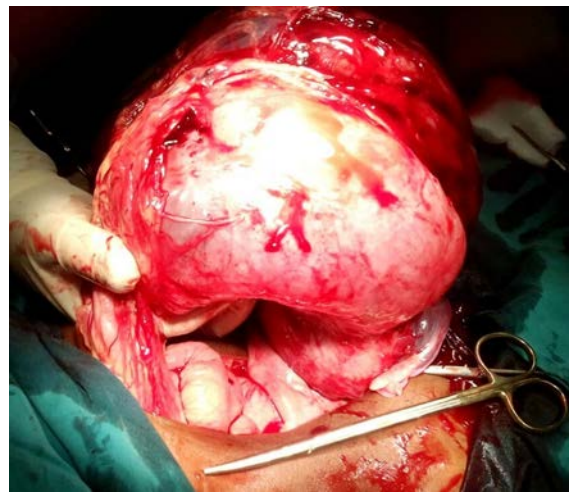
**Figure 2: Abdominal ultrasound image of solid and cystic mass**

The abdominal tomographic images revealed a 20x18x16 cm sized multiloculated mass with malignant and ovarian originated appearance, focal calcified and fat tissue density, including solid and cystic components completely filling the abdomen. Anatomic structures of the uterus and bilateral ovaries could not be clearly visualized (Figure 3). CA-125 was 1245. Laparotomy was planned for the patient with normal laboratory findings. The abdominal examination revealed a regular contoured, ovarian derived benign mass covering the left tube and ovary, which was adhered to

the omentum in its upper side (Figure 4).



**Figure 3: Abdominal tomographic image of solid and cystic mass.**

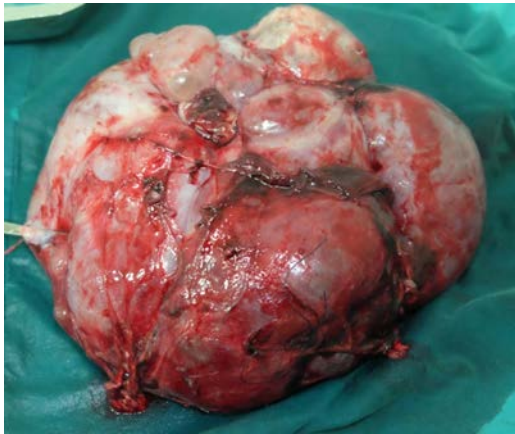


**Figure 4: Intraoperative appearance of mass.**

All organs within the abdomen were examined, however, no accompanying nodular lesion was detected except for the 0.5 cm sized small nodular lesions adjacent and adherent to the teratoma mass in the omentum. The patient underwent left salphingo-oophorectomy



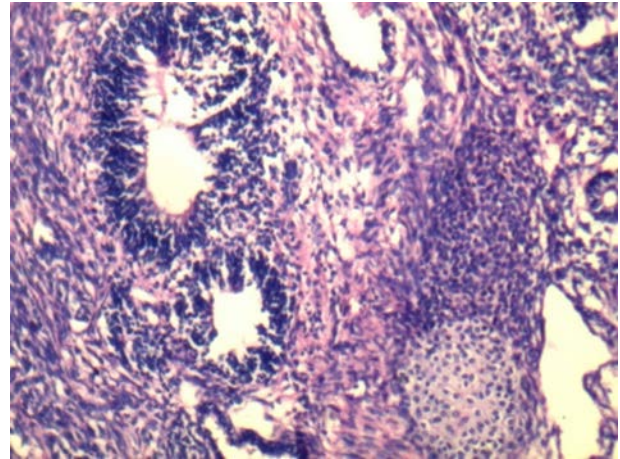
and omentectomy (Figure 5).



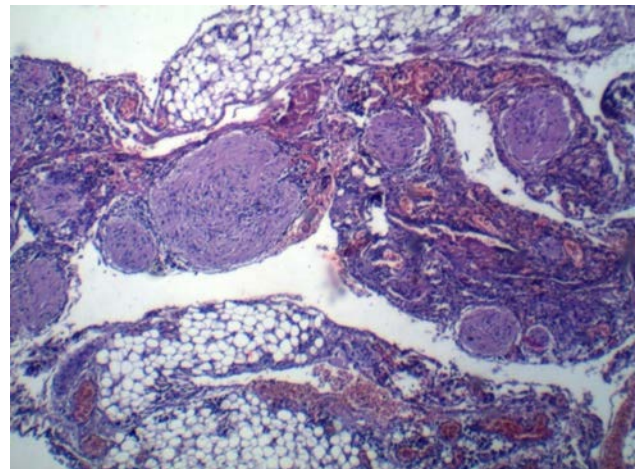
**Figure 5: Postoperative view of mass.**

The resected material was sent for pathological examination. On gross inspection, the pathological examination revealed the left ovarian mass measured 16 cm. The cut section was variegated with solid and cystic areas. Hard areas, keratinous debris and tufts of hair were also identified. Omentum, received as fibrofatty tissue, showed fine nodular deposits. Microscopically, most areas of the tumor were composed of abundant mature glial tissue along with skin and adnexal structure, glandular elements, mature and immature cartilage. Focal areas showed primitive neuroepithelium in the form of primitive neural tubes and rosettes, with rare mitosis (Figure 6). The omental implants were composed of nodular mature glial tissue (Figure 7). A diagnosis of immature teratoma (Grade 1) with multiple glial implants in omentum (Grade 0) was made. Because of a FIGO 1A and grade 1 differentiation tumor, we decided not to give chemotherapy. Sometimes, a primary site benign tumor will have a

metastatic malignant tumor as well as a primary malignant tumor may have benign metastases. Additionally, due to a concern for future reproductive fertility, the decision was made to avoid chemotherapy. No pathological finding has been detected in the 3-month follow up period of the case.



**Figure 6: Immature teratoma with neuroepithelial rosette formation (H&Ex10).**



**Figure 7: Glial implants in omental tissues (H&Ex10).**

## **Discussion**

Ovarian masses in children are different from those of adults regarding histopathological, clinical and prognostic properties, and methods of diagnosis and treatment.<sup>12</sup> Ovarian pathologies arise in a rather immature base physically, hormonally, and immunologically. Since the pelvic region is superficial and at the same time its localization is higher and closer to the median plane compared to the adult, ovarian pathologies may be diagnosed at an earlier stage and easier compared to the adults.<sup>12,13</sup>

The radiological and laboratory examinations of our 7-year-old patient with increasing abdominal swelling and pain, in addition to nausea and vomiting complaints for the last 6 months, revealed a mass located close to the median plane with both solid and cystic components that completely filled the abdomen.

Immature teratomas are lesions that may include all three germ cell layers like mature teratomas.<sup>3</sup> Immature components are commonly of mesenchymal or ectodermal origin. Immature neuroectodermal tissue is the easiest to distinguish among all other tissues. Immature teratomas are distinguished from mature teratomas by their malignant clinical progression, being observed at younger ages (commonly in the first 2 decades), being more rare and generally unilateral, and their including histologically immature or embryonic cells.<sup>4,5</sup> Patients may sometimes admit to clinics with limbic encephalitis-like rare paraneoplastic syndrome<sup>14</sup>

Immature teratomas constitute 3% of all ovarian teratomas, 1% of all ovarian cancers, and 20% of malignant germ cell ovarian tumors.<sup>3,6</sup>

The malignancy risk increases with the rate of increase of neuroepithelial tissues in immature teratomas. Peritoneal gliomatosis is a rare situation where mature glial implants are detected on the peritoneum, and the majority are immature teratoma related.<sup>15,16</sup> Gliomatosis peritonei was first defined in 1905 as the co-existence of ovary-related teratoma and mature glial tissues over the peritoneum and omentum.<sup>17</sup> Approximately 100 cases have been reported in literature by this time.<sup>18</sup>

The presence of mature glial implants that accompany immature teratomas may be considered as an indicator of good prognosis independent from the stage of the primary tumor.<sup>19</sup> The formation mechanism of peritoneal or omental neuroepithelial implants is not yet clearly defined but, two different hypothesis have been presented. According to the first hypothesis, these glial implants are genetically related to the primary tumor. It is related to the angio cephalic distribution due to the capsular defect which arises spontaneously or as a result of surgical intervention.<sup>20</sup> According to the second hypothesis—which is commonly accepted—glial cells are genetically independent from the primary tumor and arise from normal cells that originate from peritoneum or neighboring mesenchymal tissues. These cells are considered pluripotent mullerian stem cells that have introduced the metastatic process as a result of a neoplastic

stimulation with an unknown origin.<sup>21</sup>

Some of the teratoma cases with recurrence may be considered related to tumor prone syndrome. It has been believed that one or more types of peritoneal cells facilitate tumor formation as a result of exogenous or endogenous neoplastic stimuli in tumor prone syndrome.<sup>22</sup>

Glial fibrillary acidic protein (GFAP) is a protein that is produced within astrocytes of growth stage, and a dense presence in samples obtained from glial implants is an indicator of good differentiation and maturity.<sup>23</sup>

In patients with extra-ovarian involvement, the appearance of immature teratoma related glial implants gains prognostic importance. Mature glial cells detected in peritoneum or lymph nodes do not affect the prognosis negatively. These glial implants evaluated as grade 0 include mostly mature tissue. Gliomatosis peritonei is particularly observed in women with immature ovarian teratoma, but may very rarely be seen in patients who underwent ventriculoperitoneal shunt due to hydrocephalus or during pregnancy.<sup>24,25</sup>

Conservative surgery is generally the first-line treatment for immature teratomas. Chemotherapy with the surgery has been reported to affect the prognosis positively.<sup>27-29</sup> Chemotherapy is not necessary in the presence of mature glial implants in immature teratomas; however, it has been observed that concomitant chemotherapy induces the maturation of the implants in case these glial implants

are immature.<sup>27</sup> The incidence of tumor recurrence is increased with factors such as tumor stage, capsular involvement, vascular invasion, or the growth pattern of the tumor in immature teratomas.<sup>23</sup>

In the literature review of Chou et al., the prognosis of 65 cases with gliomatosis peritonei has been reported good after primary surgical treatment.<sup>15,26</sup>

In patients with immature teratoma including mature glial implants, malignant tumor lesions may be observed late after the first surgery. Therefore, long-term follow-up of these patients is recommended.<sup>7</sup>

We have followed up on our 7-year-old patient with immature teratoma and mature gliomatosis peritonei in the omental tissue after the primary surgical treatment for 3 months without any recurrence.

## **Conclusion**

Immature teratomas belong to the germ cell tumor group, frequently observed in childhood. Gliomatosis peritonei is the distribution of the neural tissue within the teratomas into the abdomen, particularly to the omentum and peritoneum. It is important that surgeons perform a post-operative clinical follow-up and help with the planning of additional treatments. They must also be sure to properly sample cases with peritoneal or omental gliomatosis of prognostic value that accompany immature teratomas.

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